Interstitial Pulmonary Fibrosis: An atypical presentation of Multiple Myeloma

Dr. Ankit Dave[1], Dr.Bikky Chaurasia[2], Dr. Hemant Joshi[3], Dr. Utsav Unadkat[4], Dr.Arti Muley[5], Dr.Nirmit Yajnik[6]

ABSTRACT

Introduction: Multiple Myeloma (MM) is a plasma cell dyscrasia, most often seen in old age.[1] Lytic bone lesion, anemia, kidney failure, hypercalcemia and recurrent infection are the most common features. [2,3] Pulmonary parenchyma is an uncommon site of involvement in multiple myeloma. [4] Interstitial lung disease is rarer; only isolated cases have been reported in the literature. [4-7] We report a case of multiple myeloma with rarely seen lung involvement in the form of interstitial pulmonary fibrosis. **Case report:** A 55/M farmer presented with generalized weakness since three months, low backache since two months and breathlessness on exertion since eight days. He had pallor and icterus but no clubbing, cyanosis or lymphadenopathy. He had bilateral scattered crepitations and tender anterior superior iliac spine and was investigated on OPD basis as well as also in the hospital during this time. However patient had atypical presentation in form of anemia, USG abdomen showed hepatomegaly and renal parenchymal disease. X ray lumbosacral spine and X ray skull revealed osteopenia and multiple lytic lesions. CXR-PA view showed reticulonodular pattern all over the lung fields. Anemia, renal involvement and lytic lesions gave rise to suspicion of multiple myeloma. Bone marrow examination showed 50% clonal plasma cells in lymphoid series and obscuration of all the three series by plasma cells. Serum electrophoresis showed elevated beta-2-microglobulin level to 3.7gm/dl (N-1.3to1.5 gm/dl). Diagnosis of MM was made based on the above findings.. On further investigation, urine was found to be positive for Bence-Jones proteins and M band. The findings of crepitations in chest associated with reticulonodular pattern of fibrosis in both lung fields on chest X-Ray PA view, suggestive of interstitial pulmonary fibrosis and hepatomegaly with icterus were unusual findings in a case of MM. **Conclusion:** Pulmonary parenchyma involvement is rare in multiple myeloma. In the few cases described, it is usually secondary to an infectious process or primary bone involvement. [8] Symptomatic liver disease is extremely rare although jaundice is occasionally seen.[9]

We are discussing this case because it showed pulmonary symptoms as the primary complaint at presentation, with simultaneous involvement of kidneys and liver.

Keywords: atypical presentation, Interstitial Pulmonary Fibrosis, Multiple Myeloma

[1, 4] 3rd year resident, [2,3] Second year resident, [5] Associate Professor, [6] Professor & head of Unit
Department of Medicine, SBKS Medical College & MIRC, piperia, Vadodara, Gujarat, India
Corresponding author mail: ankitdave_7384@yahoo.co.in

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INTRODUCTION

Multiple myeloma (MM) is a plasma cell dyscrasia, most often seen in old age. [1]. Lytic bone lesion, anemia, kidney failure, hypercalcemia and recurrent infection are the most common features. [2,3]The presence of at least 10% plasma cell in bone marrow, presence of monoclonal protein in serum or urine, and end organ damage are the diagnostic features of MM. [4,5]

Pulmonary parenchyma is an uncommon site of involvement in multiple myeloma. [6]Interstitial lung disease is rarer; only isolated cases have been reported in the literature. [6-9]One study described 13 cases with lung involvement in multiple myeloma, of which six had pneumonia, two had mass lesions, two had multiple nodular lesions, and only three had interstitial infiltrates. [9]We report a case of multiple myeloma with rarely seen lung involvement in the form of interstitial pulmonary fibrosis.

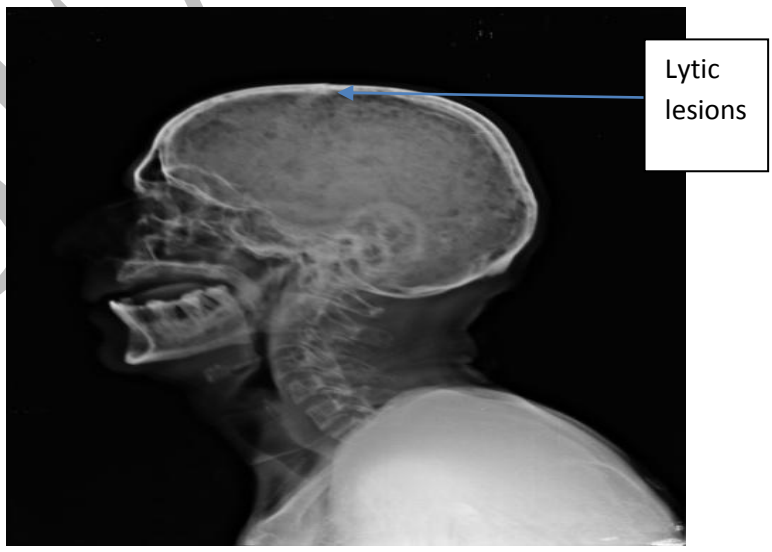
CASE REPORT

A 55-year old farmer presented with generalized weakness since three months, low backache since two months and breathlessness on exertion since eight days. His backache was of moderate intensity, gradually progressive and radiated up to mid thigh bilaterally. Initially he was able to perform his daily activities but at the time of presentation he needed support to stand and walk. Breathlessness started eight days back

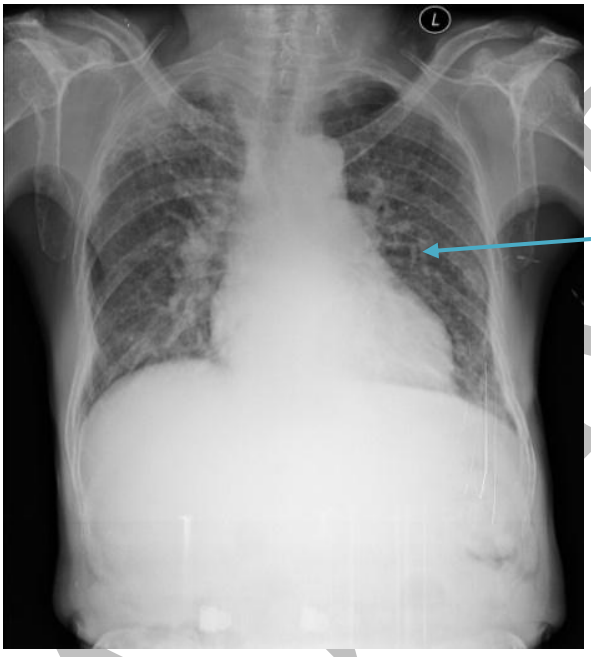
which had progressed to grade 2 NYHA at presentation.

He had a history of recurrent jaundice but no history of fever, diabetes mellitus, hypertension, ischemic heart disease or bronchial asthma. There was no significant surgical or family history. On examination, he was conscious and well oriented, with average built. His pulse was 100/minute, regular, respiratory rate was 24/minute and BP was 150/100 mm Hg. He had pallor and icterus but no clubbing, cyanosis or lymphadenopathy .He had bilateral scattered crepitations and tender anterior superior iliac spine. Rest of the systemic examination was normal.

Lab investigations revealed anemia (Hb – 5.4gm%), TLC – 8,600/cu.mm and platelet count 1.5 lakh/cu.mm. Peripheral smear was suggestive of severe hypochromic anemia with adequate platelets. His blood urea was 62, S. Creatinine - 2.3 and S. Calcium was 10.1. Urine and LFT were normal. X ray lumbosacral spine and X ray skull (Figure 1) revealed osteopenia and multiple lytic lesions.

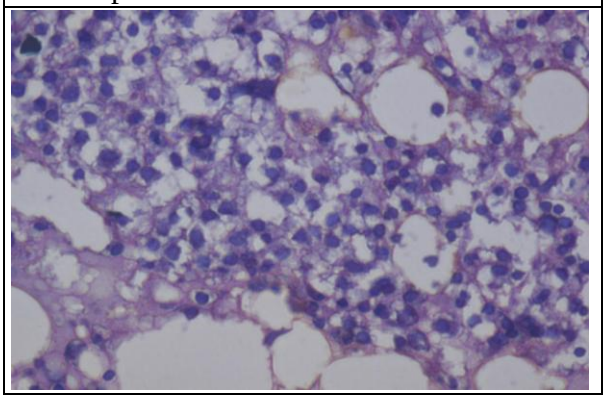


USG abdomen showed hepatomegaly and renal parenchymal disease. CXR-PA view (Figure 2) showed reticulonodular pattern all over the lung fields. Sputum AFB and gram staining were negative.ECG was normal.



Viral markers were also negative. Anemia, renal involvement and lytic lesions gave rise to suspicion of multiple myeloma. On further investigation, urine was found to be positive for Bence-Jones proteins and M band. Bone marrow examination (Figure 3) showed 50% clonal plasma cells in lymphoid series and obscuration of all the three series by plasma cells. Serum electrophoresis showed elevatedbeta-2-microglobulin level to 3.7gm/dl (N-1.3 to 1.5 gm/dl). Diagnosis of MM was made based on the above findings.

Bone marrow biopsy showing > 50% plasma cells in bone marrow.



However, the findings of crepitations in chest associated with reticulonodular pattern of fibrosis in both lung fields on chest X-Ray PA view, suggestive of interstitial pulmonary fibrosis and hepatomegaly with icterus were unusual findings in a case of MM. Hence, he needed further investigation but the patient did not give consent for bronchoscopy or liver biopsy. The patient was given symptomatic treatment with nebulization and steroids. His breathlessness was relieved to some extent with the medications and the patient was referred to higher centre for further management in view of his fast deteriorating clinical condition. However, he deteriorated fast and expired before further investigations could be done.

DISCUSSION

Among the various organs potentially affected, pulmonary parenchyma involvement is rare, and, in the few cases described, it is usually secondary to an infectious processor extending from primary bone involvement.[10] In practice however, lung involvement is so rare that lung investigations have been ignored in several MM series. [11] In a series of 958 patients with myeloma, 10% showed pulmonary findings, but only one case had plasma cell involvement demonstrated histologically, and the clinical course was suggestive of plasma cell involvement in only 3 other cases. [12]

The most common clinical manifestation of liver disease is hepatomegaly, which is not always related to plasma cell infiltration. [11,13] Symptomatic liver disease is extremely rare although jaundice is occasionally seen, almost always in association with hepatic amyloid deposits. [13]

Our case of MM had involvement of lung as interstitial pulmonary fibrosis and hepatomegaly with jaundice, which are

rarely seen with MM. Another finding which was atypical was that despite wide spread lytic lesions, he did not have hypercalcemia. However, the diagnosis of extramedullary pulmonary dissemination of MM cannot be established as the patient did not give consent for bronchoscopy and broncho alveolar lavage or a lung biopsy.

CONCLUSION

We are discussing this case because it showed a rare presentation of MM, simultaneous involvement of kidneys, lungs and liver along with anemia and widely spread lytic lesions but normal calcium levels and pulmonary symptoms as the primary complaint at presentation. His clinical condition deteriorated fast, especially once the respiratory complaints started appearing. This case brings to our notice that plasma cell neoplasm should be kept in the differential diagnosis of pulmonary infiltrate not responding to antibiotics as it may be a marker of rapidly progressing disease.

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